Chapter

Audio-Vestibular Neurosensory Prosthetics: Origins, Expanding Indications and Future Directions

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Abstract

Approximately one-third of persons over 65 years are affected by disabling hearing loss. It is estimated that the number of people with disabling hearing loss will grow to 630 million by 2030 and maybe over 900 million by 2050. Deafness has significant consequences on many aspects of an individual's life, including their socioeconomic status, mental and physical well-being, educational and employment opportunities. When congenital or early in the developmental years, deafness results in a delay or loss of language acquisition. Deafness can result from damage or disease anywhere along the auditory pathway. Hearing prosthetic devices help restore hearing and the use of these devices depends on the degree and type of hearing loss. This chapter will give a brief account of the currently available prosthetic hearing solutions.

Keywords: auditory brainstem implant, auditory midbrain implant, bone conduction implant, cochlear implant, hearing aid, middle ear implant

1. Introduction: the neurosensory problem

The global burden of disabling hearing impairment is estimated at 466 million people (6.1% of the world's population) where 432 million (93%) of these are adults (242 million males, 190 million females) and 34 million (7%) children. It is estimated that the number of people with deafness will grow to 630 million by 2030 and maybe over 900 million by 2050 [1].

Hearing impairment has a significant bearing on many aspects of an individual's life, including their socioeconomic status, mental well-being, education and employment opportunities. Older people with moderate or more severe hearing loss were more likely to feel depressed and suffer with poor mental health [2]. The deaf child cannot listen to her or his mother and focus on an activity simultaneously since both inputs must be processed visually. In addition, the deaf child is unaware of sounds of the outside environment, and thus, is centered on self and own activities. This has consequences on the child's development of language, social skills and cognition [3].

Hearing impairment results from damage or disease anywhere along the auditory pathway. Surgical restoration of hearing involves procedures that range from ossicular reconstruction to implantation of devices which serve to assist the functioning of the auditory pathway. The prosthetic devices currently being used for restoration of hearing are classified based on their mode of action in **Figure 1**.

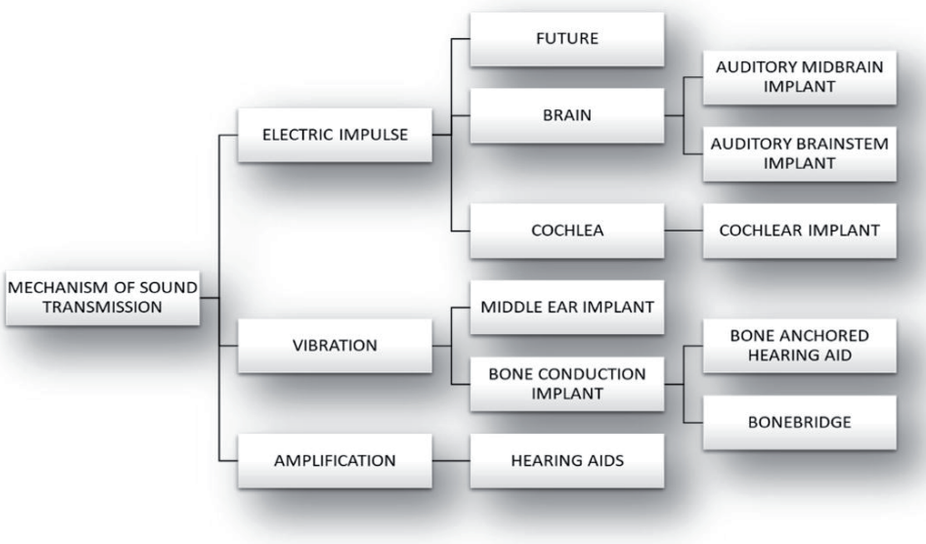

Figure 1.
Classification of hearing prosthetic devices.

2. The auditory prosthetic solutions

The first prosthetic used for the management of deafness on record in human history was the *Ear Trumpet or Horn*. These were funnel shaped devices which collected sound waves and led them into the ear, thus strengthening the impact of sound energy on the ear drum and improving hearing. The use of these devices date back to the 17th century with the earliest description given by the French Jesuit priest and mathematician Jean Leurechon in his work "*Recreations Mathematiques*" (1634). Commercial production of these devices began much later in the 1800s and many notable personalities of that era including pianist Ludwig van Beethoven are known to have used them.

2.1 Candidacy assessment

Hearing impaired individuals usually seek help only when they have reached a stage that they can no longer ignore their hearing loss. Often, it is not the individual themselves, but the family member/caretaker who notes that the concerned individual is struggling with their hearing impairment. In either situation, the pre-audiometric assessment which involves collection of relevant medical history and clinical examination of the hearing-impaired person is an important first step of the auditory rehabilitation process. It is important to identify patients in whom medical or non-prosthetic surgical management of their hearing loss should be attempted prior to dispensing of the hearing device. After the clinical assessment, the candidacy process involves a complete audiological evaluation using the following tests.

Pure Tone Audiometry (PTA) – This is a subjective test involving hearing threshold evaluation in both air conduction and bone conduction.

Speech Audiometry – This test provides information concerning hearing for speech, the type and degree of hearing impairment, and to check the reliability of the pure tone thresholds.

Brainstem Evoked Response Audiometry (BERA) – This test is an objective and non-invasive method of hearing assessment which detects electrical activity along the auditory pathway (from inner ear to inferior colliculus) in response to sound impulse.

Oto-Acoustic Emissions (OAE) – This involves recording of a low-level sound emitted by the cochlea either spontaneously or evoked by an auditory stimulus thus indicating integrity of the outer hair cells of the cochlea.

Cortical Auditory Evoked Potentials (CAEP) – This test is an objective and non-invasive assessment of electrical activity from the level of the inferior colliculus to the primary auditory cortex in response to sound stimulus.

Aided Audiometry – This test is similar to PTA but involving the estimation of hearing thresholds during the use of a hearing prosthesis.

2.2 The hearing aid (HA)

2.2.1 History

The simplest and most widely used auditory prosthetic solution is the Hearing Aid. The first electronic hearing aids in the mid-19th century were large instruments that sat on a table. The subsequent invention of the transistor and miniature electron tubes made the hearing aid small enough to fit behind the ear. In the late 1960s the introduction of minicomputers opened the doors to real-time signal processing for people with hearing loss. Although not fast enough, these were computer-controlled analog systems which were used for amplification in hearing aid devices. In 1975, Daniel Graupe developed the first digital hearing aid in which high speed digital-array processors were used. This made it possible to process audio signals digitally in real time. Finally, in the early 1980s, the Central Institute of the Deaf (CID) developed the first practical wearable digital hearing aid which set precedent to the current type of aids in use [4].

2.2.2 Design

The modern digital hearing aid is a marvel of sophisticated engineering and miniaturization. The microphone is the first component to receive the sound signal and it converts this energy into electricity. These microphones may be unidirectional, bidirectional or omnidirectional. A more sophisticated type of an omnidirectional microphone is an "adaptive directional microphone" which works together with the hearing aid's noise reduction technology to highlight speech sounds and important environmental sounds while suppressing background noise. These small signals generated by the microphone are sent to the amplifier which makes them more powerful. Compression amplifiers are used to amplify the signal while avoiding distortion and decreasing its dynamic range and can represent sound in either an analogue (mimicking acoustic waveforms) or digital (representing signals as a string of numbers) manner. The signal then goes through a filter which is used to change the relative amplitude of the high, mid and low frequency characteristics of a signal. Thus, the sound signal can be altered by the user or clinician to suit the type of hearing loss. Finally, the receiver converts this modified and amplified electrical signal back into sound with the help of electromagnetism which is similar to how headphones work. The hearing aid performs all these functions with the help of electrical power from a detachable battery. Based on where they are worn, hearing aids are classified as body (pocket aids), spectacle, behind-the-ear (BTE), in-the-ear (ITE), in-the-canal (ITC) and completely-in-canal (CIC). BTE are further classified based on location of the receiver as receiver-in-the-aid (RITA) where the receiver lies within the hearing aid case or receiver-in-the-ear (RITE) where the receiver lies within the ear canal.

2.2.3 Mode of action

The primary mode of action of a hearing aid involves amplification of the input sound signal accompanied by frequency specific sound signal modulation to produce a tailormade output sound signal aimed at supporting the targeted hearing impairment. Hearing aid technology has seen rapid growth in the past 50 years and these advances can be broadly classified into three generations, all of which are currently commercially available. The first generation were simple analog hearing aids which were adjusted with screwdriver-controlled potentiometer trimmers according to the degree of hearing loss. An analog hearing aid consists of a microphone, preamplifier, a tone controller (or automatic gain controller - AGC), an amplifier and a receiver. In such devices, an acoustic signal is converted by the microphone into an electric signal which is amplified by a preamplifier and the frequency is shaped by the tone controller. This signal is then again amplified and converted into an acoustic output signal by the receiver. These were followed by second generation analog devices which could be digitally programmed by dedicated devices or computers. This involved coding of certain parameters of the analog components such as tone control and allowing these settings to be stored in a memory that can be modified or retrieved according to user preference. Digital hearing aids, which represent the third-generation, differ from the analog devices in that the amplified electronic signals are converted into digital signals which are processed by the Digital Signal Processor (DSP) before being converted back to analog electronic signals. Thus, these devices usually have an analog-to-digital converter, a digital signal processor and a digital-to-analog converter, of which the latter function is often embedded within the receiver in the newer devices. The entire system is built on a single integrated circuit that contains all the electronic parts i.e. transistors, capacitors and resistors [5].

2.2.4 Contralateral routing of signal (CROS)/binaural CROS (BiCROS)

CROS are hearing aids worn bilaterally where the hearing aid on the affected side transmits the acoustic signal to the receiver in a hearing aid worn on the better hearing ear. In patients who have hearing loss in the better hearing ear, the hearing aid in the better ear can also be used to provide amplification to that ear in addition to the CROS input. This configuration is called BiCROS [6].

2.3 The bone conduction device (BCD)

2.3.1 History

In the 1st century, Pliny the Elder, a Roman scientist, was the first to remark about the potential of sound conduction through solid bodies. Several centuries later, Cardano demonstrated a method by which sound may be transmitted to the ear by means of the shaft of a spear held between one's teeth [7]. About a hundred years later, in the 1600s, it was Hieronymus Capivacci, an Italian physician, who realized the clinical significance of Cardano's observations. Using the same experiment he determined that if the patient heard the sound it indicated disease of the tympanic membrane, while if the patient could not hear the sound it indicated a lesion of the auditory nerve [8]. In 1757, Johann Jorissen, a German physician, published the first known dissertation dealing exclusively with hearing through teeth. But it was only in 1821, when the first primitive bone conduction device was developed by Jean-Marie Gaspard Itard, a French ear specialist who postulated that sound was conducted through the bones of the entire skull [9]. Using the concepts

of the carbon microphone and the magnetic receiver (earphone) in 1920, Joseph Prenn of Boston, patented a mechanical bone-conductive ear [10]. However, it was Frederick Kranz of Illinois, who in 1925, patented the first real bone conduction vibrator which was handheld initially, and later attached to a headband and set the tone for bone conduction technology [11]. With the help of the Sonotone Company, Hugo Lieber developed a small wearable bone conduction receiver in 1933 [12]. Another idea was to fix the bone conduction device in eye glasses. From the 1960s to the 1990s, four different companies – Amplivox, Akumed, Otarion & Oticon made bone conduction eyeglasses which became the most widely used bone conduction hearing devices of that time. The idea to implant the vibrator into the mastoid bone originated in Sweden, after the pioneering work of the anatomist, Per-Ingvar Brånemark in bone rheology, where Anders Tjellström fitted the first 3 patients with the BAHA implant in 1977 [13].

2.3.2 Design, mode of action & candidacy

Bone conduction devices are prosthetic devices that aid hearing by converting sound energy into vibrational energy. The vibrations that are produced are transmitted to the skull bone, and then to the inner ear bypassing a hearing impairment in the external or middle ear, thus overcoming the air conduction defect. Below is a basic classification of bone conduction devices (BCD) based on their mode of action: *Direct-drive* (sound vibration is sent to bone directly in the presence of a skin defect), *Skin-drive* (where sound vibration is sent to bone via intact skin) & *In-the-mouth* (where sound vibration is sent to the teeth). BCDs can also be classified as *non-implantable* (conventional bone conduction devices) and *semi-implantable* devices (where some part of the device is implanted).

The **Conventional Skin-Drive Non-Implantable BCD**s usually consist of a processor attached to soft headbands (softbands), steel spring headbands or spectacles. Common drawbacks of these devices are that a high static skin pressure is required to transmit the vibrations to the cochlea, leaving the skin compressed for prolonged periods, which might lead to discomfort and skin problems. Also, skin attenuates the high-frequency sound signal and therefore sound that reaches the cochlea has a lower content of high frequencies. These devices are commonly used to confirm candidacy before formal implantation of a BCD. Alternatively, they are used when surgery is contraindicated or refused. Candidacy requirements for this type of BCD include a permanent conductive hearing loss secondary to microtia, atresia or syndromic; bilateral mixed hearing loss with bone conduction average of 35 dB or less and single sided deafness where bone conduction average is 15 dB or less in the better ear. There are no age restrictions. Below are some of the common conventional skin-drive non implantable BCDs available.

2.3.3 The Cochlear BAHA SoundArc

This is a conventional skin-drive non-implantable BCD that is designed to be worn behind the head with the BAHA sound processor attached to the connector disc just behind the ear.

2.3.4 The Softband with BAHA processor attached

This is a conventional skin-drive non-implantable BCD that consists of a bone conduction processor fitted on a soft headband and is indicated in the pediatric population with hearing loss as part of their pre-implant assessment till formal surgery.

2.3.5 The Med-El ADHEAR

This is a conventional skin-drive non-implantable BCD consisting of an adhesive adapter which is placed behind the pinna on the mastoid skin and a bone conduction processor which gets attached to that adapter.

The semi-implantable BCDs have several advantages over the conventional BCDs which include a lack of static skin compression and preservation of the high frequency signals. These BCDs are further classified as *direct-drive percutaneous* devices, *direct-drive active transcutaneous* devices (with an implanted transducer) and *skin-drive passive transcutaneous* devices (with an implanted magnet).

The **Semi-implantable Direct-drive Percutaneous BCD**s involve use of a biocompatible osseointegrated implant to which a percutaneous abutment is fixed. The sound processor is then clicked on to the abutment to activate hearing. The major benefit of these devices is an efficient percutaneous transmission of sound vibrations providing maximum amplification. Candidacy requirements for this type of BCD include a conductive hearing loss with an air bone gap of more than 30 dB; mixed hearing loss with a conductive component more than 30 dB and sensorineural component of up to 65 dB HL and single sided deafness where bone conduction average is 15 dB or less in the better ear. Suitable for ages 5 years and above.

2.3.6 The Cochlear BAHA CONNECT

The BAHA CONNECT system was the first commercially available bone conduction device. This direct-drive percutaneous system consists of a titanium implant which is osseointegrated into the skull, along with a sound processor which transmits these vibrations through a percutaneous abutment which connects the sound processor to the implant. The skin is left intact around the abutment using *'Dermalock'* technology. The Cochlear BAHA 5 sound processor features the *'BCDrive'* electromagnetic transducer. This transducer creates vibrations that are sent through to the cochlea via an abutment in the case of BAHA CONNECT, or an implanted and an external magnet in the case of BAHA ATTRACT (described later). This sound processor can also be used in the conventional way by connecting to either the Softband or the SoundArc. The BAHA 5 System consists of a range of sound processors including the BAHA 5 Sound Processor (for up to 45 dB SNHL), the BAHA 5 *Power* Sound Processor (for up to 55 dB SNHL) and the BAHA *SuperPower* Sound Processor (for up to 65 dB SNHL) which address different levels of hearing loss.

2.3.7 The Oticon PONTO 4

The Oticon PONTO 4 system is a direct-drive percutaneous device that consists of three parts – a 4 mm titanium implant that is surgically implanted into the skull, an abutment that is seamlessly placed percutaneously through the skin and a sound processor that clicks easily to the abutment and sits discreetly behind the ear. In addition to its remarkably small size, the PONTO 4 system also connects wirelessly to the internet using an IFTTT network.

The **Semi-implantable Skin-drive Passive Transcutaneous BCD**s have a major advantage in complete elimination of the soft tissue & skin complications of a percutaneous abutment but their disadvantages include the large artifact area on post-implant MRI scans and the possible dampening of high-frequency sound signals through the skin. Candidacy requirements for this type of BCD are similar to the direct drive percutaneous BCDs.

2.3.8 The BAHA ATTRACT

The BAHA ATTRACT system is a skin-drive passive transcutaneous device that transmits sound vibrations to the inner ear through a magnetic connection between the sound processor and the implant. The magnet which lies on the inside to the skin is attached to the underlying skull bone with a screw, and the BAHA sound processor is attached to a magnet plate on the skin via a soft pad to equalize the force distribution over the attachment surface. This BCD offers the benefit that there is no skin penetrating abutment, thus providing a good esthetic outcome with no need for daily care.

2.3.9 Medtronic ALPHA 2 MPO

The Medtronic ALPHA 2 MPO system is a skin-drive passive transcutaneous device that consists of a surgically implanted internal plate containing two airtight sealed magnets and the external digital sound processor coupled to the base plate containing twin magnets corresponding to the internal ones. In order to overcome the skin problems related to high static skin pressure, it uses a larger surface area so that the static force is widely distributed alleviating dermal compression [14].

In **Semi-implantable Direct-drive Active Transcutaneous BCD**s, the transducer is implanted in bone under intact skin. Hence, vibrations are transmitted directly from the transducer to the skull bone with the elimination of dermal impedance. These BCDs are labeled transcutaneous since it is an electromagnetic signal from the sound processor that is transmitted through the skin to the implanted transducer and not sound vibrations. Candidacy requirements for this type of BCD are similar to the direct drive percutaneous devices (except for age 12 years and above for Cochlear OSIA).

2.3.10 The Med-El BONEBRIDGE

The BONEBRIDGE is a direct-drive active transcutaneous system that consists of an external audio processor and an internal bone conduction implant. The internal component has a receiver coil, a magnet, a demodulator, and a cylindrically shaped bone-conduction floating mass transducer (BC-FMT) secured to the bone by two titanium screws. The power to drive the FMT is transmitted transcutaneously to the internal coil, processed by the demodulator and then relayed to the BC-FMT, which then transduces the signals into mechanical energy. Osseointegration of the titanium screws, however, is not thought to be crucial.

2.3.11 The Cochlear OSIA

The OSIA System is a direct-drive active transcutaneous system that uses a Piezo-Power transducer which sits within the OSI200 implant, and is positioned under the skin to send sound to the cochlea. The OSI200 implant is positioned on top of the bone and connected to the osseointegrated BI300 Implant which gives an important single-point transmission for sound to the skull. The system has a fitting range of 55 dB SNHL. The transducer functions on the principle of the "piezoelectric effect" which is the ability of certain materials to generate vibrations when provided with an electrical charge.

2.3.12 In-the-mouth BCD: SoundBite

This in-the-mouth BCD is neither direct-drive nor skin-drive. The vibrations are generated by a piezoelectric transducer and are transmitted through the teeth

to the skull. SoundBite by Sonitus was mainly developed for single sided deafness patients. A microphone is placed behind the deaf ear and sound is sent wirelessly to an in-the-mouth transducer transmitting vibrations to the upper molar teeth. These vibrations are transmitted to the skull bone and received by the healthy cochlea. This device is currently only available for investigational use for the management of single sided deafness and is not available commercially [15].

2.4 The active middle ear implant (AMEI)

2.4.1 History

Alvar Wilska, a Finnish physiologist, is credited with the first attempt at mechanical stimulation of the auditory system with the help of an electromagnetic driver. In 1935, he sprinkled iron filings on to the tympanic membrane of a patient lying on his side and placed a ear-phone over the man's ear that produced no sound, but an electromagnetic signal, and the patient reported hearing. Subsequently, in 1959, Rutschmann devised a method of fixing a tiny permanent magnet to the tympanic membrane at the umbo with water soluble glue. By introducing an alternating current he produced pure tones in the range of 2 kHz to 10 kHz [16]. In 1973, Goode and Glattke refined this work by introducing an electromagnetic coil on the postauricular skin to drive the magnet fixed on the umbo. Heide reported an important modification to these previous studies in 1988, by replacing the postauricular transducer with an in-the-canal electromagnetic induction coil located millimeters from a magnet fixed at the umbo [17]. Prior to this in 1986, Maniglia at Case Western Reserve University had already begun investigating one of the first contactless electromagnetic based middle ear implant systems. In that same year, Kartush and Tos, at the Michigan Ear Institute, collaborated with Smith & Nephew Richards, to create an electromagnetic-based partially implantable middle ear device that used an in-the-canal electromagnetic coil with a custom ear mold housing [18]. The use of piezoelectric crystals in middle ear implants became evident in 1984, when the RION device became the first commercially approved piezoelectric-based middle ear device to be implanted. Since this time, several additional middle ear implants have used this technology, including the Impex TICA and the Envoy Esteem.
This was roughly a century after Piezoelectricity was first discovered by Jacques and Pierre Curie in 1880 after observing that certain solid substrates develop an electrical charge proportional to an applied mechanical stress. In 1996, Geoffrey Ball pioneered development of the VIBRANT SOUNDBRIDGE which became the first FDA approved Active Middle Ear Implant (AMEI) system for implantation in patients with SNHL, receiving approval in August 2000 [19].

2.4.2 Design, mode of action & candidacy

These can be classified as *totally implantable* if all these parts are beneath the skin or *partially implantable* if only the receptor and transducer are implanted beneath the skin. Depending on the type of energy the transducer utilizes, MEI are classified as: *Piezoelectric* – when energy is transmitted to a piezoelectric crystal, which deforms it or changes its volume generating a vibratory signal; *Electromagnetic* – when energy is transmitted to a coil that generates an electromagnetic field that causes a magnet located close to the ossicular chain or the inner ear to vibrate and thus produce a vibratory signal; or; *Electromechanical* – which are similar to the electromagnetic implants but the coil and magnet are closely related to each other and the ossicular chain [20]. The main advantages of the AMEI over hearing aids is that they induce direct mechanical vibration of the ossicular chain or

(and) the intracochlear fluids and thus generate greater functional gain, especially over the acute frequencies, with less distortion and feedback, increasing spoken-word discrimination. In terms of subjective results, patients report better intelligibility and quality of sound and a more natural perception of their own voice than obtained with hearing aids [21].

2.4.3 The Med-El VIBRANT SOUNDBRIDGE

The VIBRANT SOUNDBRIDGE (VSB) is currently the most widely implanted middle ear device worldwide. This AMEI consists of an externally worn processor that contains the microphone, electronic signal processor and battery and an implantable part called the Vibrating Ossicular Prosthesis (VORP). This VORP contains a magnet (that enables the external part to be coupled), a receiver unit, a demodulator (that filters the signal received), a conductor link for the electrical signal and a floating mass transducer (FMT). Sound signal received by the external audio processor is transmitted transcutaneously to the implanted device generating vibratory movements of the FMT and conduction of sound to the inner ear. The FMT coupled to the Incus is the original indication devised for patients with a healthy middle ear and moderate–severe sensorineural hearing loss in the higher frequencies with a discrimination >50% at conversational intensity who were not satisfied with their hearing aid or who had repeated external otitis. The VSB is the only AMEI approved for use in children since 2009. It is indicated in children with external auditory canal atresia and bilateral malformations of the ossicular chain who have sensorineural conductive or mixed hearing loss and meet the same audiological criteria as for adults.

2.4.4 The Cochlear CARINA

This is a totally implantable AMEI which consists of two parts, the electronic capsule that contains the microphone, the batteries, the digital processor and the connector, and the middle ear transducer that contains the receiver unit and the electromechanical transducer. All these parts are placed beneath the skin with no need for externally worn processors. This AMEI is indicated for adults with moderate to severe SNHL with a hearing threshold between 30 and 85dBHL especially in the higher frequencies.

2.4.5 CODACS (direct acoustic Cochlear implant)

The CODACS implant is a semi-implantable device that is still in a preliminary phase and is gradually being used in some European centers. This AMEI is indicated for patients with severe to profound mixed hearing loss due to otosclerosis either as a primary indication or after stapedial surgery has failed.

2.4.6 The envoy ESTEEM implant

The Envoy ESTEEM implant is a fully implantable device which is indicated in patients with moderate to severe and severe sensorineural hearing loss. The system uses two piezoelectric transducers (PZTs). Sound is received via a PZT sensor that picks up eardrum vibrations and transforms them into an electric signal. This signal is filtered, modified, amplified and transferred to a PZT driver which mechanically drives the stapes thus conducting sound to the inner ear. The sound processor also contains a power source, which is an implantable lithium iodide battery [22]. For candidacy, hearing thresholds should be stable and between 35 and 85dBHL for

audiometric frequencies of 500-4000 Hz with a word recognition score of 40% or greater. It is currently indicated only in patients older than 18 years.

2.5 The Cochlear implant (CI)

2.5.1 History

The history of the cochlear implant dates back to the first attempts at electrical hearing. The Italian scientist Alessandro Volta (1800) was the first to demonstrate that electric stimulation could directly evoke auditory sensations in humans when he invented the battery [23]. However electric stimulation of the auditory system was not subsequently reported for another 150 years until modern electronic technology appeared. SS Stevens and his colleagues conducted a series of studies to re-examine the electric stimulation of hearing using vacuum based tube oscillators and an amplifier, a copper wire serving as an electrode. They identified three mechanisms that were responsible - the first mechanism was called an "electromechanical effect" by Kiang & Moxon in 1972 in which electrical stimulation causes the hair cells in the cochlear to vibrate, resulting in a perceived tonal pitch at the signal frequency it was acoustically stimulated; the second mechanism occurs due to the tympanic membrane's conversion of the electric signal into an acoustic signal, resulting in a tonal pitch perception but at the doubled signal frequency; while, the third mechanism is due to direct electric activation of the auditory nerve. However, it was Andreev who first gave direct evidence of electric stimulation of the auditory nerve when hearing sensations were reported with electric stimulation in a deaf patient whose middle and inner ears were damaged [24]. The modern era of cochlear implants began when Djourno and Eyries successfully performed the electric stimulation of hearing in two deafened patients in 1957 [25]. Their success led to a frenzied increase in attempts to restore hearing to deaf people on the US west coast in the 1960 and 1970s. Although their methods were crude, these studies identified critical problems and limitations that needed to be considered and overcome for successful implementation of electric hearing. In 1984, the House 3 M single-electrode implant became the first Food and Drug Administration (FDA) approved device. This was followed by the Ineraid or Symbion device developed by the University of Utah, the Laura device developed by the University of Antwerp, and the Digisonix MX20 developed by the MXM laboratories in France. These devices were later phased out and are no longer commercially available [26]. At present, there are three major cochlear implant manufacturers including Med-El Corporation, Austria; Cochlear Corporation, Australia and Advanced Bionics, USA. In addition to its design, implantation criteria have evolved over the past decades. Niparko provides a detailed account of these evolving criteria in patient selection as well as the surgical, cost-utility, educational, pre and post-operative issues in cochlear implants [27]. Most notably, the audiological criteria for cochlear implantation has relaxed from bilateral total deafness (<110 dB HL) in the early 1980s to severe hearing loss (>70db HL) in the 1990s, and then to current suprathreshold speech based criteria (<50% open-set sentence recognition with properly fitted hearing aids).

2.5.2 Design, mode of action & candidacy

The essential components of a cochlear implant are as follows – a microphone converts sound into an electrical signal for input to the speech processor. The processor transforms this electrical input into a set of stimuli for the implanted array of electrodes. These stimuli are sent to the electrodes via a transcutaneous link which

involves encoding of the stimulus information for efficient radiofrequency transmission from an external transmitting coil to an internal (implanted) receiving coil. The signal received by the internal coil is decoded to specify the electrical stimuli for the electrode array. These electrical stimuli that stimulate the hair cells have signal characteristics which determine the sound quality of the perceived stimulus.

2.5.3 Med-El SYNCHRONY CI

The SYNCHRONY cochlear implant system consists of the SYNCHRONY 2 cochlear implant with the Sonnet 2 BTE or Rondo 2 sound processor. The implanted device has a removable magnet that may be temporarily taken out by a cochlear implant surgeon in the event that the MRI (up to 3.0 Tesla) is needed for the head. Med-El offers the largest selection of electrode options within the cochlear implant industry. Traditional electrode options include the standard array, medium array, compressed array and split electrodes array. The newest options include the FLEX 24, 28 and FLEX SOFT electrodes. The Sonnet 2 is a redesigned BTE sound processor for greater ease of use, better esthetics and improved reliability. It recognizes the ambient environment or scene and automatically adjusts settings to match it. The Rondo 2 is a unique sound processor option different from the Sonnet 2. It has innovative wireless charging, simple on/off button and automatically controls volume level for the recipient with up to 18 hours of battery life. The device is small and compact and sits on the head, just behind and above the ear and more comfortable to wear with glasses as compared to any BTE system.

2.5.4 The advanced bionics NAIDA CI

The HiRes Ultra 3D cochlear implant has a multi-magnet assembly which automatically provides alignment to the 3D MRI field, allowing adult and pediatric users to undergo 3.0 Tesla MRIs safely, without any preparation surgery or head bandaging. This unique magnet assembly is composed of four rotatable magnets encased in a revolving disc allowing alignment with the 3D MRI field. There is no need for head bandaging or possible surgical removal of the magnet. The electrode array options in Advanced Bionics include the HiFocus Mid-Scala electrode array – which is designed to lie in the center of the scala tympani in order to remain reasonable proximal to the modiolus (and cochlear neural elements) while not making physical contact with the delicate cochlear structures necessary to maintain residual hearing; and, the HiFocus Slim J electrode array – which is designed to be placed in the scala tympani toward the lateral cochlear wall away from the cochlear structures. The Naída CI Connect is a design-integrated solution which turns the Naída CI Q90 sound processor into a Bluetooth headset which allows for hands-free calling and direct audio streaming from any compatible device.

2.5.5 Cochlear nucleus PROFILE PLUS CI

Built on the same design of the predecessor Profile series, Profile Plus provides easier access to MRI at 1.5 Tesla and 3.0 Tesla without the need for compression or magnet removal. The Profile Plus is available with the following electrode types: Slim Modiolar (CI632), Slim Straight (CI622), Slim 20 (C624) & Contour Advance (CI612). The Contour advance electrode array has a perimodiolar design and a soft tip which is designed to reduce trauma to the delicate cochlear structures during insertion using the Advanced Off-Stylet (AOS) technique. The Straight array is recommended for patients with cochlear anomalies like the common cavity and in situations where there is fibrous tissue growth in the cochlea (bacterial meningitis).

The Cochlear NUCLEUS 7 Sound Processor is the smallest and lightest BTE hearing solution with built-in connectivity featuring direct streaming with a compatible Apple and Android device without attaching anything to the sound processor. The Kanso 2 Sound Processor is the smallest and lightest off-the ear solution available that has built-in technology offering direct streaming, control and connectivity with a compatible device.

2.5.6 Candidacy

Current clinical guidelines for implant candidacy represent a composite assessment of the individual's age (>12 months), hearing loss history, aided and unaided audition (comprehensive audiometric assessment), the socioeconomic and family support of the patient likely to influence the use of the device; and an awareness of potential benefits and constraints of current implantable technologies. These can be estimated via high resolution imaging complemented by thorough audiometric assessment and a multidisciplinary team-based preoperative counseling or intervention.

2.6 The auditory brainstem implant (ABI)

2.6.1 History

Hitselberger and House performed the first implantation of an ABI in 1979 [28]. It was a single spherical electrode implanted adjacent to the cochlear nucleus following removal of bilateral vestibular schwannomas of a patient with Neurofibromatosis Type 2 (NF2). In 1992, Cochlear Ltd. in conjunction with the Huntington Medical Research Institute developed the first multichannel electrode array devices which have, in most cases, provided patients with greater benefits than the earlier devices [29].

2.6.2 Design, mode of action & candidacy

Although in the earlier years the most frequent indication of an ABI was NF2, a genetic disorder with an autosomal dominant inheritance pattern in which the patient develops benign vestibular schwannomas on both eighth cranial nerves; in recent years, the indications for the ABI have expanded to include bilateral cochlear ossification, unilateral vestibular schwannoma with deafness in the contralateral ear that is not amenable to a cochlear implant, congenital cochlear nerve aplasia or hypoplasia, complete cochlear ossification, malformation of the inner ear, and bilateral traumatic avulsions or absence of the cochlear nerve [30]. The current ABI system utilizes 12 platinum disk electrodes aligned on a flexible silicone and polyester mesh backing for implantation into the lateral recess of the fourth ventricle on the cochlear nucleus of the brain. This bypasses a non-functioning auditory nerve and allows signals to be sent to the brain.

2.7 The auditory midbrain implant (AMI)

2.7.1 History

The first reported attempt at stimulation of the inferior colliculus (IC) of the midbrain for hearing restoration was in 1962 by Simmons and colleagues at Stanford University during a tumor removal operation [31]. The second attempt at stimulating the surface of the IC happened much later in 2005 and involved using

an ABI array in a NF2 patient to assess if the limited performance with cochlear nucleus stimulation secondary to tumor related damage could be overcome by stimulation in a higher auditory center and they were successful in eliciting a response at much higher thresholds than what is used in cochlear nucleus stimulation [32]. In 2006, Thomas and Minoo Lenarz at the Hannover Medical University developed the first human prototype Auditory Midbrain Implant (AMI) array for penetrating stimulation across the tonotopic gradient of the central nucleus of the IC [33].

2.7.2 Design, mode of action & candidacy

The AMI is a type of central auditory prosthesis that targets midbrain regions beyond the cochlear nucleus, particularly the central nucleus of the Inferior Colliculus (ICC). There are several properties of the ICC that make it a logical choice for a prosthetic target including being a converging center for almost all ascending auditory projections, possessing tonotopic anatomical organization in addition to spatial organization for speech perception. The Cochlear Ltd. CI array was reduced in dimensions to create an AMI array that was small enough to insert into the ICC with the goal of stimulating its different layers. The AMI electrode array measures 6.4 mm long and a diameter of 0.4 mm and possesses 20 linearly spaced platinum ring electrodes. A distal Dacron mesh prevents over insertion of the electrode into the ICC during implantation and anchors the electrode array onto the surface of the inferior colliculus to minimize movement after positioning. The other components of the AMI system are similar to the Cochlear NUCLEUS CI system consisting of behind-the-ear microphone and processor that transmits the electromagnetic signals to the receiver stimulator implanted under the skin.

2.8 The vestibular implant

2.8.1 History

Vestibular disorders are widely prevalent and can cause significant morbidity in the form of incapacitating symptoms, missed work days and even the inability to leave one's home. Advancements in the treatment of vestibular disorders are lacking and the surgical treatment of vestibular disorders has remained unchanged in the past two decades. Over the same duration, neural prosthesis have been developed that substitute one modality for another in order to restore a missing sense. Current research in vestibular neurostimulation has finally made possible the clinical reality of treating a wide range of vestibular disorders with electric stimulation.

2.8.2 Design, mode of action & candidacy

Vestibular implantation is useful in recurrent acute vertigo attacks (in which central compensation does not occur), for chronic bilateral hypofunction and for Meniere's disease, in which vertiginous attacks may be precipitated by an acute loss of vestibular tone. These are ideal candidates for a "pacemaker"-style vestibular implant that replaces the missing neural impulses during attacks. This implant bypasses the vestibular end organs to directly stimulate the vestibular nerve. The goal of vestibular implantation is to provide vestibular functionality and or reduce symptomatology by programmed stimulation of the vestibular nerve. This device consists of a modified Nucleus Freedom system. The receiver-stimulator has a modified trifurcating array of 9 electrodes. Each electrode is implanted 2.5 mm into the perilymphatic space of the semicircular canal adjacent to the ampullary nerve which is the site of stimulation.

3. Future directions

It would be expected that future devices will meet the existing basic requirements which include wearing comfort, cosmetic appeal, long battery life and customizable frequency-cum-level dependent gains for ideal hearing rehabilitation. There is extensive ongoing research in different aspects of hearing technology, some of which are described here. The micro-electrical–mechanical systems (MEMS) microphone allows for a further reduction in size as well as the possibility of multiple microphones on a single device. This will be very useful for noisy environments for selecting a target source while rejecting other competing sounds thus improving directionality of the hearing device [34]. Silicon microfabrication of microphones making them more sensitive and with lower thermal noise was inspired by the ears of the parasitoid fly, *Ormia ochracea* and is another unique approach [35]. As the size of the microphone progressively reduces, self-calibrating devices are likely to become more common in the future. Such systems can help in achieving target frequency-and-level dependent gains at the initial fitting and can greatly speed up the initial fitting process. Initiation of the hearing device could itself trigger a self-adjustment procedure to ensure the device overcomes daily variations [36]. DSPs have progressively shrunk in size and power requirement which together with improvements in battery technology will contribute to increased intervals between battery recharging. Developments in both battery chemistry and internal components such has anodes will likely result in longer battery life, faster recharging, smaller size and increased voltage that will enable increased dynamic range and DSP processor speed [36].

Bluetooth is one such technology that hearing device manufacturers are increasingly making available in their devices. This makes the hearing device compatible with any Bluetooth enabled technology across manufacturers allowing for hands-free usage of phone and other sound devices. Future hearing devices may contain sensors already existent in smart watches and other wearable devices which may collect information and present it via an auditory speech signal tailored to the wearer. There is evidence that auditory evoked electrical responses change depending on which sound source an individual is attending to [37]. This has been referred to as cognitively controlled hearing aids and research is ongoing in this field at present. The possibilities for improving the bone conduction device in the future are endless. A better housing design and improved transducer technology can improve conventional BCDs. This would be a significant advantage since they are non-invasive and a lower-cost alternative. Expanding surgical criteria to children below a certain age with smaller devices would likely offer them better hearing at a younger age and improved socio-linguistic development. Development of more patient optimized and powerful transducers will expand current indications for implantation.

A variety of approaches to regeneration or repair of auditory sensory and related structures are being investigated to restore hearing. Future hearing devices may be developed consisting of an acoustic hearing aid and a linked implanted component capable of eluting chemicals or signals for enhancement of this regenerative process. Drug delivery would be enabled through the implant, to preserve neurons or even promote the growth of neurites (toward the electrode array) from existing neurons or regeneration of neurons and associated structures [38, 39]. In 2002, the Free Electron Laser was used to conduct initial experiments on optical stimulation of a peripheral nerve [40]. This concept was later transferred to the cochlea and stimulation units have been further miniaturized and an implantable unit for stimulation in cat models is currently under work [41]. Necessary safety studies are underway that has led to the development of an optogenetic implant unit that

is being used in clinical trials. The feasibility of supplementing the ABI array of surface electrodes with penetrating microstimulating electrodes has been demonstrated in animal studies and human trials involving using this combination is currently ongoing. The initial results of the first AMI patients have been encouraging in terms of the ability to implant the array into the auditory midbrain safely and to restore some hearing function. Future work in this area will needed to overcome the primary limitation of midbrain stimulation which is optimal placement of the electrode array. Research is also currently underway on creating a totally implantable vestibular implant in which all the components of the implant could be internalized. Advances in miniaturization, microelectromechanical systems (MEMS), nanotechnology, battery technology and circuit energy efficiency could help realize this goal. A vestibular brainstem implant could prove helpful for bilateral vestibular hypofunction and such a device could be potentially combined with an auditory brainstem device [29].

Thus, there are new development efforts in this field that will either significantly improve prosthetic performance or change the face of auditory prosthesis altogether. There is also the possibility that auditory prostheses will be integrated with other peripheral and central prostheses (eg. vestibular and deep brain implants) to treat not just one symptom but to address its whole spectrum. Finally, the progress in neuroscience, particularly in non-invasive brain monitoring will allow a full account of individual variability in both the optimization & the performance of prosthetic hearing devices.

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References

[1] Davis AC, Hoffman HJ. Hearing loss: Rising prevalence and impact. *Bull World Health Organ*. Published online 2019. doi:10.2471/BLT.19.224683

[2] MI W, WJ S, SJ S, J K, GA K. Comparative impact of hearing and vision impairment on subsequent functioning. *J Am Geriatr Soc*. Published online 2001.

[3] URBAN E. Childhood Deafness: Compensatory deintegration of the self. *J Anal Psychol*. Published online 1989. doi:10.1111/j.1465-5922.1989.00143.x

[4] Levitt H. Digital hearing aids: Wheelbarrows to ear inserts. *ASHA Lead*. Published online 2007. doi:10.1044/leader.ftr4.12172007.28

[5] Holube I VTM. *Textbook of Hearing Aid Amplification*. 2nd ed. (Sandlin R E, ed.). Singular; 2000.

[6] Snapp H. Nonsurgical Management of Single-Sided Deafness: Contralateral Routing of Signal. *J Neurol Surgery, Part B Skull Base*. Published online 2019. doi:10.1055/s-0039-1677687

[7] Parisiis F& R. *Cardano G: De Subtilitate Libri XXI.*; 1550.

[8] Johannis, Beyeri H. *Capivacci H: Opera Omnia Cura*. Francofurti, Paltheniena; 1603.

[9] JMG I. *Traité Des Des Maladies de l'oreille et de l'audition*.; 1821.

[10] Prenn J. *Mechanical Bone-Conductive Ear*. US Patent 1346721

[11] Frederick Kranz. *Vibration Instrument for Bone Audition*. US Patent 1540783

[12] Hugo Lieber. *Hearing- Aid Device*. US Patent 1940553

[13] Tjellstrom A, Lindstrom J, Hallen O, Albrektsson T, Brånemark PI. Osseointegrated titanium implants in the temporal bone. A clinical study on bone-anchored hearing aids. *Am J Otol*. Published online 1981.

[14] Bernardeschi D, Russo FY, Nguyen Y, et al. Audiological Results and Quality of Life of Sophono Alpha 2 Transcutaneous Bone-Anchored Implant Users in Single-Sided Deafness. *Audiol Neurotol*. Published online 2016. doi:10.1159/000445344

[15] Hkansson B. The future of bone conduction hearing devices. *Adv Otorhinolaryngol*. Published online 2011. doi:10.1159/000323715

[16] Rutschmann J. Magnetic Audition—Auditory Stimulation by Means, of Alternating Magnetic Fields Acting on a, Permanent Magnet Fixed to the Eardrum. *IRE Trans Med Electron*. Published online 1959. doi:10.1109/IRET-ME.1959.5007893

[17] Heide H, Tatge G ST. Development of a semi-implantable hearing device. In: Suzuki I; Hoke M, ed. *Middle Ear Implant: Implantable Hearing Aids*. Basel: Karger; 1988:32-43.

[18] Kartush JM, Tos M. Electromagnetic ossicular augmentation device. *Otolaryngol Clin North Am*. Published online 1995. doi:10.1016/s0030-6665(20)30574-0

[19] Ball GR. The Vibrant Soundbridge: Design and development. *Adv Otorhinolaryngol*. Published online 2010. doi:10.1159/000318516

[20] Beleites T, Neudert M, Bornitz M, Zahnert T. Sound transfer of active middle ear implants. Otolaryngol Clin North Am. 2014;47(6):859-891. doi:10.1016/j.otc.2014.08.001

[21] Tysome JR, Moorthy R, Lee A, Jiang D, Fitzgerald O'Connor A. Systematic review of middle ear implants: Do they improve hearing as much as conventional hearing aids? *Otol Neurotol*. Published online 2010. doi:10.1097/MAO.0b013e3181db716c

[22] Maurer J, Savvas E. The Esteem system: A totally implantable hearing device. *Adv Otorhinolaryngol*. Published online 2010. doi:10.1159/000318523

[23] Marchese-Ragona R, Pendolino AL, Mudry A, Martini A. The Father of the Electrical Stimulation of the Ear. *Otol Neurotol*. Published online 2019. doi:10.1097/MAO.0000000000002153

[24] Andreev AM, Gersuni GV VA. On the electrical excitability of the human ear: on the effect of alternating currents on the affected auditory apparatus. *J Physiol USSR.* 1935;(18):250-265.

[25] Eisen MD. Djourno, eyries, and the first implanted electrical neural stimulator to restore hearing. *Otol Neurotol*. Published online 2003. doi:10.1097/00129492-200305000-00025

[26] Zeng FG, Shannon R V. Loudness-coding mechanisms inferred from electric stimulation of the human auditory system. *Science (80-)*. Published online 1994. doi:10.1126/science.8160013

[27] Francis HW, Niparko JK. Cochlear implantation update. *Pediatr Clin North Am*. Published online 2003. doi:10.1016/S0031-3955(03)00034-8

[28] Edgerton BJ, House WF, Hitselberger W. Hearing by cochlear nucleus stimulation in humans. *Ann Otol Rhinol Laryngol*. Published online 1982.

[29] Schwartz MS, Otto SR, Shannon R V., Hitselberger WE, Brackmann DE. Auditory Brainstem Implants. *Neurotherapeutics*. Published online 2008. doi:10.1016/j.nurt.2007.10.068

[30] Colletti V, Carner M, Miorelli V, Guida M, Colletti L, Fiorino F. Auditory brainstem implant (ABI): New frontiers in adults and children. *Otolaryngol - Head Neck Surg*. Published online 2005. doi:10.1016/j.otohns.2005.03.022

[31] Simmons FB, Mongeon CJ, Lewis WR, Huntington DA. Electrical Stimulation of the acoustical nerve and the inferior colliculus. *Arch Otolaryngol*. Published online 1964. doi:10.1001/archotol.79.6.559

[32] Colletti V, Shannon R, Carner M, et al. The first successful case of hearing produced by electrical stimulation of the human midbrain. *Otol Neurotol*. Published online 2007. doi:10.1097/01.mao.0000247808.47712.02

[33] Lenarz T, Lim HH, Reuter G, Patrick JF, Lenarz M. The auditory midbrain implant: A new auditory prosthesis for neural deafness-concept and device description. *Otol Neurotol*. Published online 2006. doi:10.1097/01.mao.0000232010.01116.e9

[34] Lewis BJ, Moss B. MEMS Microphones, the Future for Hearing Aids. Analog Dialogue. 2013;47(11):3-5.

[35] Miles RN, Hoy RR. The development of a biologically-inspired directional microphone for hearing aids. Audiol Neurotol. 2005;11(2):86-94. doi:10.1159/000090681

[36] Popelka GR, Moore BCJ. Future Directions for Hearing Aid Development. Published online 2016:323-333. doi:10.1007/978-3-319-33036-5_11

[37] Mesgarani N, Chang EF. Selective cortical representation of attended speaker in multi-talker speech

perception. *Nature*. Published online 2012. doi:10.1038/nature11020

[38] Clark GM. Cochlear implants: climbing new mountains The Graham Fraser Memorial Lecture 2001. *Cochlear Implants Int*. Published online 2001. doi:10.1179/cim.2001.2.2.75

[39] Qun LX, Pirvola U, Saarma M, Ylikoski I J. Neurotrophic factors in the auditory periphery. In: *Annals of the New York Academy of Sciences*. ; 1999. doi:10.1111/j.1749-6632.1999.tb08649.x

[40] Wells J, Kao C, Konrad P, et al. Biophysical mechanisms of transient optical stimulation of peripheral nerve. *Biophys J.* Published online 2007. doi:10.1529/biophysj.107.104786

[41] Cao Z, Xu Y, Tan X, Richter C-P, Suematsu N, Robinson AM. Electrical and optical co-stimulation in the deaf white cat. In: ; 2018. doi:10.1117/12.2291839